

WHAT IS CLAIMED IS:

- Sub B1
- 1 ~~1. A method of treating a patient with Pompe's disease, comprising:~~
2 administering to the patient a therapeutically effective amount of human acid alpha
3 glucosidase.
 - 1 ~~2. The method of claim 1, wherein the patient is administered at least 10~~
2 mg/kg body weight per week.
 - 1 3. The method of claim 1, wherein the patient is administered at least 60
2 mg/kg body weight per week.
 - 1 4. The method of claim 1, wherein the patient is administered at least
2 120 mg/kg body weight per week.
 - 1 5. The method of any of claims 1-4, wherein the patient is administered a
2 single dosage of alpha-glucosidase per week.
 - 1 6. The method of any of claim 1-4, wherein the patient is administered
2 three dosages of alpha-glucosidase per week.
 - 1 7. The method of any of claims 1-4, wherein the amount is administered
2 per week for a period of at least 24 weeks.
 - 1 8. The method of claim 1, wherein the alpha-glucosidase is administered
2 intravenously.
 - 1 9. The method of claim 1, wherein the alpha-glucosidase was produced
2 in milk of a transgenic mammal.
 - 1 10. The method of claim 1, wherein the patient has infantile Pompe's
2 disease.
 - 1 11. The method of claim 10, wherein the patient survives to be at least
2 one year old.
 - 1 ~~12. The method of claim 1, wherein the patient has juvenile Pompe's~~
2 disease.

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1 ~~13. The method of claim 1, wherein the patient has adult Pompe's disease.~~

1 14. The method of claim 1, wherein the alpha-glucosidase is
2 predominantly in a 110 kD form.

1 15. The method of claim 1, further comprising monitoring a level of
2 human acid alpha glucosidase in the patient.

1 16. The method of claim 15, further comprising administering a second
2 dosage of human acid alpha glucosidase if the level of alpha-glucosidase falls below a
3 threshold value in the patient.

1 17. The method of claim 1, wherein the human alpha glucosidase is
2 administered intravenously and the rate of administration increases during the period of
3 administration.

1 18. The method of claim 17, wherein the rate of administration increases
2 by at least a factor of ten during the period of administration.

1 19. The method of claim 17, wherein the rate of administration increases
2 by at least a factor of ten within a period of five hours.

1 20. The method of claim 17, wherein the patient is administered a series
2 of at least four dosages, each dosage at a higher strength than the previous dosage.

1 21. The method of claim 20, wherein the dosages are a first dosage of
2 0.03-3 mg/kg/hr, a second dosage of 0.3-12 mg/kg/hr, a third dosage of 1-30 mg/kg/hr and a
3 fourth dosage of 2-60 mg/kg/hr.

1 22. The method of claim 21, wherein the dosages are a first dosage of 0.1-
2 1 mg/kg/hr, a second dosage of 1-4 mg/kg/hr, a third dosage of 3-10 mg/kg/hr and a fourth
3 dosage of 6-20 mg/kg/hr.

1 23. The method of claim 22, wherein the dosages are a first dosage of
2 0.25-4 mg/kg/hr, a second dosage of 0.9-1.4 mg/kg/hr, a third dosage of 3.6-5.7 mg/kg/hr
3 and a fourth dosage of 7.2-11.3 mg/kg/hr.

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1 24. The method of claim 23, wherein the dosages are a first dosage of
2 0.3 mg/kg/hr, a second dosage of 1 mg/kg/hr, a third dosage of 4 mg/kg/hr and a fourth
3 dosage of 12 mg/kg/hr.

1 25. The method of any of claims 20-24, wherein the first, second, third
2 and fourth dosages are each administered for periods of 15 min to 8 hours.

1 26. The method of any of claims 20-24, wherein the first, second, third
2 and fourth dosages are administered for periods of 1 hr, 1hr, 0.5 hr and 3 hr respectively.

1 27. A pharmaceutical composition comprising human acid alpha
2 .glucosidase, human serum albumin, and a sugar in a physiologically acceptable buffer in
3 sterile form.

1 28. The pharmaceutical composition of claim 17 comprising human
2 acid alpha glucosidase, human serum albumin, and glucose in sodium phosphate buffer.

1 29. A pharmaceutical composition comprising alpha glucosidase,
2 mannitol and sucrose in an aqueous solution.

1 30. The pharmaceutical composition of claim 27, wherein the sugar
2 comprises mannitol and sucrose and the concentration of mannitol is 1-3% w/w of the
3 aqueous solution and the concentration of sucrose is 0.1 to 1% w/w of the aqueous
4 solution.

1 31. The pharmaceutical composition of claim 27, wherein the
2 concentration of mannitol is 2% w/w and the concentration of sucrose is 0.5% w/w.

1 32. A lyophilized composition produced by lyophilizing a
2 pharmaceutical composition comprising human acid glucosidase, mannitol and sucrose in
3 aqueous solution.

1 33. A pharmaceutical composition prepared by
2 lyophilizing a first composition comprising human acid alpha-
3 glucosidase, mannitol, sucrose and an aqueous solution to produce a second composition;
4 and reconstituting the lyophilized composition in saline to produce a third composition.

SUB A1

add B2

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